Unusual Presentation of Posteriolateral Congenital Diaphragmatic Hernia

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with very unusual presentation. One of the three had a presentation which has not been reported in the medical literature to the best of our knowledge. This infant presented with acute peritonitis at the age of two months. Laparotomy confirmed the diagnosis of strangulated stomach in posterolateral diaphragmatic hernia.

POSTERIOLATERAL congenital diaphragmatic hernia is one of the most serious causes of respiratory distress in neonates and infants. The age of the neonate at the time of the clinical onset of the symptom is very important. Presence of respiratory distress within the first twenty-four hours in an ominous sign. This suggests that the ipsilateral lung is hypoplastic, while the neonates who present with respiratory distress 24 hours after birth have a very good chance of complete recovery following surgical intervention. Most of the neonates who have an ipsilateral hypoplastic lung do very well in immediate post-operative period but unfortunately deteriorate after few days of a honey-moon period. This starts with hypoxia and carbon dioxide retention and ends in cardiopulmonary arrest. These very unfortunate events are a result of complicated vascular and biochemical changes due to intrapulmonary arterio venous shunting and pulmonary hypertension.

The condition of the ipsilateral lung tissue depends on the timing of the intra-abdominal contents herniation into the thorax. If this herniation occurs in the first trimester, the herniated intraperitoneal contents will compress the developing lung and result in a very hypoplastic lung. If the herniation occurs in the last few months of pregnancy or after birth, the lung would have already developed and the herniated intraperitoneal contents will not affect its development but it compresses the ipsilateral lung, causing mediastinal shift to the other side and compressing the contralateral lung tissue. During the past two years six cases of congenital posteriolateral diaphragmatic hernia were detected at Salmaniya Medical Center. There were three cases

CASE NO. 1

A two months old Bahraini male infant, presented with vomiting of twenty-four hours duration. The vomiting occured few minutes after feeding and was not bile stained. Three hours before admission the abdomen became distended, and he became hypoactive. Twelve hours before admission his parents took him to a doctor for treatment.

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The baby was a product of term pregnancy with normal delivery. His birth weight was 3.2 Kg. He is the first child in the family. His father is a Bahraini and his mother an Egyption. He was very healthy before admission and never had any respiratory or gastrointestinal problems before.

Physical examination showed a well nourished baby. He was toxic, in shock with poor capillary filling. His pulse rate was 200/minute, respiratory rate was 82/minute, temperature 102°C and blood pressure was not recordable. The abdomen was severely distended and tympanic with generalized tenderness and rebound. Bowel sounds were not audible. The chest was clear with good air entry on both sides. The abdominal x-rays showed a large amount of intraperitoneal gas under the diaphragm with multiple fluid levels all over the abdomen, suggesting intraperitoneal perforation with diffuse peritonitis. The supine chest x-ray showed a rounded soft shadow mass in the left lower lung field. (Fig. 1). The lateral view confirmed its position in the lower posterior part of the left chest. The erect chest x-ray showed an elevated left diaphragm to the level of 5th intercostal space and there was gas with a soft tissue mass under it. (Fig. 2). The haematological investigation showed haemoglobin concentration of 8.9 gm/100 ml; hematocrit 30%, white count 24000 with 74% neutrophils. The electrolyte report showed : Sodium 124 mmol/100 ml; potassium 6.8 mmol/100 ml; chloride 84 mmol/100 ml, Bicarbonate 8 mmol/100 ml; Urea 48 mmol/100 ml and Serum Creatinine 2 mg/100 ml.

After resuscitation, fluid and electrolyte replacement and starting of Gentamicin 5 mg/Kg with Keflin 100 mg/Kg, Laparotomy

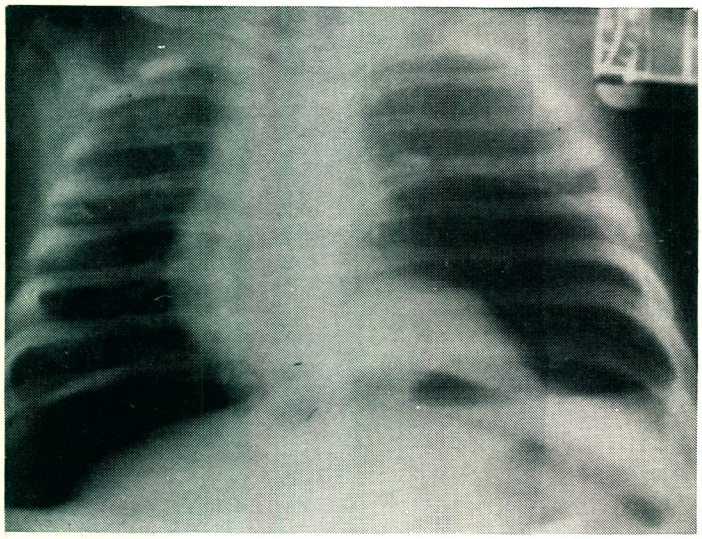

FIG. 1 *Chest and erect abdominal x-ray of Case No. 1 – shows elevation of the diaphragm with gas under it and a soft shadow mass in the left side.*

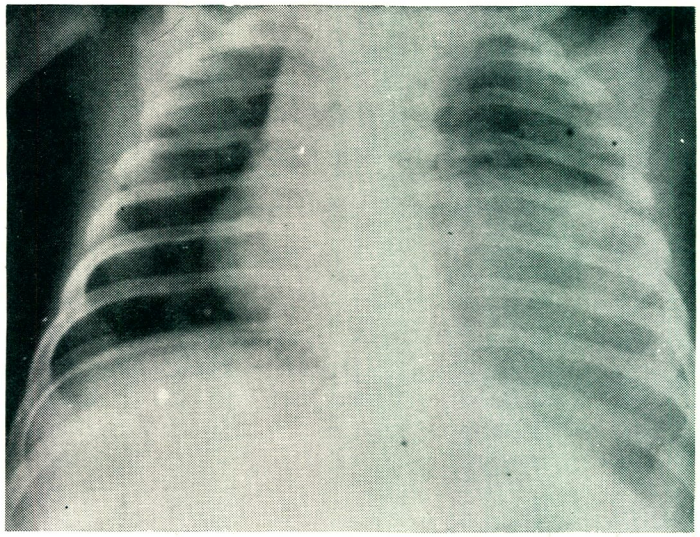

FIG. 2 *Supine chest and abdomen x-ray of Case No. 1 – shows a soft shadow mass in the left lower hemithorax.*

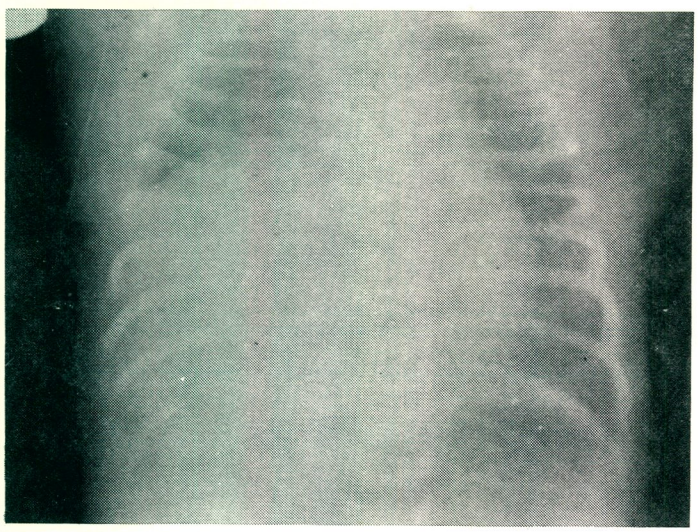

FIG. 3 *Chest x-ray of Case No. 2 – shows elevation of the right diaphragm and compression of the right lung.*

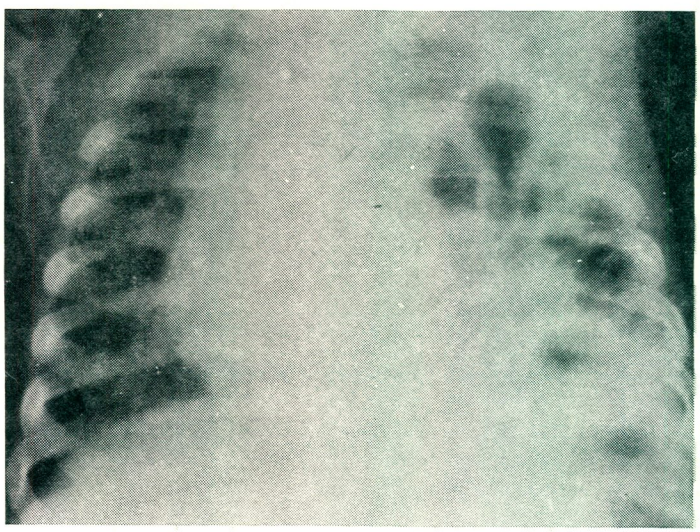

FIG. 4 *Chest x-ray of Case No. 3 – shows multiple cystic lesion in the left hemithorax with mediastinal shift to the right.*

was performed under general anaesthesia. The peritoneal cavity was filled with pus, there was a posterolateral diaphragmatic hernia with strangulated stomach. The stomach was reduced, the greater curvature was gangrenous with a large gastric perforation. Partial gastrectomy was performed including all the gangrenous and ischemic part of the left side of body and fundus of the stomach. The peritoneal cavity cleaned of the pus and washed thoroughly with warm normal saline. The hernial sac was resected, a chest tube was inserted and the posteriolateral congenital diaphragmatic opening was closed with interrupted neurolon 3/0. Two corrugated drains were inserted, one in the pelvis and the other under the diaphragm. The peritoneal cavity was closed with Dexon and the abdominal wound mopped with Bethadine solution and closed in layers with Dexon. The skin was closed with subcutaneous 4/0 Dexon. The patient had an uneventful post-operative recovery. Feeding was started on the fourth day. The chest drain was removed on the third post-operative day and the peritoneal drains on the fifth post-operative day.

CASE NO. 2

A one day old Indian male presented with tachypnea few hours after birth. He was a product of full term pregnancy with normal delivery. His birth weight was 3.6 Kg. He was the first child in the family. Physical examination showed a well nourished baby with tachypnea and mild sub-costal retraction. The chest was clear with good air entry. Chest x-ray showed moderate elevation of the right diaphragm and the lateral view (Fig. 3) showed elevation of the diaphragm with very minimal constriction at the level of the normal position of the diaphragm which made us suspect diaphragmatic hernia with a large defect and hernial sac. Fluroscopic screening of the diaphragm showed paradoxical movement which was suggestive of eventration of the right diaphragm. With oxygen therapy his tachypnea improved. Two weeks later his tachypnea recurred and there was no changes on his chest x-ray. Right posterolateral thoracotomy was performed through the sixth intercostal space. There was a very large posterolateral congenital diaphragmatic hernia with an intact sac containing all the liver. The hernial content were reduced, the hernial sac was resected and the posterolateral diaphragmatic defect closed with difficulty after inserting a chest drain. The baby had an uneventful post operative recovery. We have been following him up for the last seven months. He is thriving well and has no respiratory problems.

CASE NO. 3

A one year old Bahraini female was admitted to the Pediatric ward with history of vomiting and lose motions for twenty four hours. She was admitted one week previously with the same complaints and the only positive investigation was detection of giardia in the stool.

She was a product of full term pregnancy with normal delivery. The day after admission she started coughing and chest examination revealed few rales on the left side. Chest x-ray showed an area of pneumonitis in the left side. A Broad spectrum antibiotic was started. On the third day after admission she became severely dyspneic and cyanotic. There was some degrees of dullness by percussion on the left side of chest, and air entry was poor on the same side. Chest x-ray showed multiple cystic lesions in the left side of the chest with mediastinal shift to the right (Fig. 4). A limited Barium study was done to confirm the diagnosis of congenital diaphragmatic hernia and to rule out the suspicion of pneumatocele.

The Barium meal and follow through showed the stomach and the proximal intestines to be in the abdomen. The Barium study was not helpful but the clinical impression was that we were dealing with a case of congenital diaphragmatic hernia. A left thoracotomy was performed through the sixth intercostal space. There was a posteriolateral congenital diaphragmatic hernia with herniation of the small bowel and the spleen into the chest. There was no hernial sac. The small bowel and the spleen were reduced and the hernial defect was easily closed. The infant had an uneventful post-operative recovery. He has been followed up for the last nineteen months. He is thriving well with no respiratory problems.

DISCUSSION

In 1848 Bochdalek (1) described the embryology of diaphragmatic malformation and the posteriolateral foramen which still carries his name. During the first thirty years of this century the treatment of congenital diaphragmatic hernia was conservative (4) and the mortality was very high. In 1940 Surgery was generally accepted after Ladd and Gross (b) published their successful series.

The incidence of congenital posteriolateral diaphragmatic hernia varies from one center to another. It is extremely difficult to estimate the true incidence of this condition. First, because many of these babies will die before establishing the diagnosis and secondly because in some, the herniation of the intraperitoneal content will occur at a later period of the infants

life as in our third case (3). Bonham et al reported on 34 cases of congenital lobar diaphragmatic hernia admitted to Great Ormond Street Hospital for sick children over a period of 14 years (2). Gross reported 35 cases from Boston Children's Hospital over the same period (5). We diagnosed six cases over a period of two years. This is very large a number for a population of 350,000 and an annual birth rate of about 10,000 in Bahrain.

The most common presentation is cyanosis at birth or few hours later. Rickhame reported (8) 84 cases from Liverpool Regional Hospital during 1954 to 1975 of which 72 were cyanosed after birth. 47 of these had dyspnea and four had apnea at birth, all of whom died. Vomiting was an uncommon symptom presented only in six cases. Vomiting might indicate malrotation of the midgut or rupture of the stomach. Strangulation and gangrenous stomach is extremely rare in this condition (7). Among our cases, three neonate presented with severe dyspnea and cyanosis at birth, which is the most common and usual presentation. The other three cases had a very unusual presentation. One presented with acute peritonitis with gangrenous perforated stomach. A second one presented with mild gastrointestinal symptoms on two occasions with mild left penumonitis but ended by sudden herniation of some of the intraperitoneal content to the chest through a congenital defect which was there since birth. The explanation of the sudden herniation at one year of age is not very clear. But a sudden increase in the negative intrapleural pressure as in cases of chest infection with dyspnea can suck the abdominal contents into the chest. Another explanation might be that increase in intra-abdominal pressure can push the intraperitoneal contents into the chest through the congenital defect in the diaphragm. The third case presented with tachypnea at birth but there was some difficulty in differentiating it from diaphragmatic eventration. We should mention that we did suspect a large congenital diaphragmatic hernia in the lateral views but this was disproved by screening the diaphragm. We delayed the operation for two reasons : First, because the baby improved with oxygen therapy in the first few days; secondly we felt that if the case was an eventration needing surgery it would be safer to wait few weeks and have reasonable diaphragmatic tissue for plication.

We believe the abdominal approach is ideal for left sided hernia. It is easy to reduce the hernial content, examine the peritoneal cavity for any other congenital anomalies and avoid the post operative problems of thoracotomy. We prefer the thoracic approach for the right sided hernias which is rare. The hernial sac might be absent but if it is intact it must be resected and the hernial defect closed with interrupted non-absorable sutures. It is usually advised to close the diaphragmatic defect in two layers by overlaping we do not feel that this was necessary in our experience. A one layer simple closure is adequate easy and less time consuming in critically ill neonates under anaesthesia and furthermore in some patients there is not enough diaphragmatic muscle tissue for overlaping. It is usually not difficult to approximate the two rims of the diaphragmatic defect. In one case we had to bring the anterior rim of the diaphragmatic defect to the posterior intercostal muscles. The post-operative management is the most challenging aspect of Bochdalek's hernia, particularly if associated with lung hypoplasia. These neonates do very well in the immediate post-operative period till the honey-moon period is over After four to five days they get progressive hypoxia with carbon dioxide retention as a result of pulmonary arterio venous shunting, pulmonary vasoconstriction and hypertension. Different pulmonary vasodilators have been tried with no definite success. Ligation of patent ductus arteriosus has been attempted to decrease the amount of arterio venous shunting but with no success. The result of surgical intervention in neonates is excellent if the respiratory distress appears after the first twenty-four hours of the neonates life. This usually suggests that there is no lung hypoplasia and if there is it will be minimal. In our three reported cases all had their symptoms after the first twenty four hours of birth. One at two days of age, one at two months and the third at one year. All had uneventful post-operative recoveries inspite of the fact that case No. 1 had severe diffused peritonitis with septicemic shock and perforated gangrenous stomach.

SUMMARY

Three unusual presentation of congenital posteriolateral Bochdaleks diaphragmatic hernia were described. The first case presented at two months of age with acute peritonitis and with perforated gangrenous stomach. The second case presented with tachypnea after birth and the chest x-ray suggested eventration of the right diaphragm and the third case had a congenital posterolateral diaphragmatic defect without herniation until one year, when she presented with gastrointestinal symptom and left side penumonitis and suddenly became dyspneic, as a result herniation of intraperitoneal contents into her chest. The clinical presentation, the radiological find-

ings of these cases were described and the surgical management and the results were discussed.

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REFERENCES

1. Bochdalek, V. (1848) "Einige Betrachtungen ueber die Einstellung des angeborene Zwerchfellbruches als Beitrag zur pathologischen Anatomie der Hernien" Vietelijahrs. prakt. Heilk., 19, 89.

2. Bonham Carter, R.E., Waterson, D.J. and Aberdeen, E. (1962) "Hernia Eventration of the Diaphragm in Childhood", Lancet, i, 656.

3. Day, B. (1972) "Late appearance of Bochdalek Hernia", BR. MED. J. 1, 786.

4. Greenwald, H.M. and Steiner, M. 1929 "Diaphragmatic Hernia in Infancy and Childhood", AM J DIS Child, 38, 361.

5. Gross, R.E. (1953). The Surgery of Infancy and Childhood. Philadelphia W.B. Saunders.

6. Ladd, W.E. and Gross, R.F. (1940). "Congenital Diaphragmatic Hernia". New Engl. J MED 223, 917.

7. Rickham, P.P. (1955) "Strungulated Diaphragmatic Hernia in Newborn Period" Thorax, 10, 104.

8. Rickham, P.P., Lister J. Irving, I.M. Neonatal Surgery (1978) Butterworths London, Boston, 165. □□